Watch Dermatomyositis Patients for Cancers and Organ Involvement

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Vienna. — Adul t patients who are diagnosed with dermatomyositis should have a thorough work-up for malignancy because they are at heightened risk for various cancers, Dr. Ralph M. Trueb said at the 16th Congress of the European Academy of Dermatology and Venereology.

About 15%-30% of patients with this inflammatory autoimmune disorder will develop cancer, with the risk being most prominent early in the course of disease. Most reported malignancies are carcinomas rather than sarcomas or lymphomas, said Dr. Trueb of the University Hospital of Zurich.

The work-up should include a chest radiograph, Pap smear, and breast and rectal examination, and should be repeated at regular intervals, he said.

Patients are also at risk for potentially life-threatening organ involvement. The lung is involved in 40% of these patients, with the development of aspiration pneumonitis, interstitial fibrosis, and respiratory insufficiency. The joints, gastrointestinal tract, and cardiovascular system also can be affected. More than one-fourth of patients have arthritis, and electrocardiogram abnormalities are present in half of patients.

The condition also may be limited to the skin and muscles. The skin lesions typical of dermatomyositis can be classified as pathognomonic, characteristic, or compatible, Dr. Trueb said.

Pathognomonic findings include Gottron’s papules, which are slightly elevated violaceous papules located over the joints of the hands, as well as over the ankles, knees, and elbows, and Gottron’s sign, which refers to the erythematous plaques that spare the interphalangeal spaces.

It’s important to note that, while the skin lesions in dermatomyositis can resemble those seen in lupus, they are located over the joints in dermatomyositis and primarily between the joints in lupus,” he said.

Characteristic skin findings include a heliotrope periorbital rash with or without associated periorbital edema; the “shawl sign,” which is a symmetrical macular violaceous erythema at the nape of the neck and around the shoulders; and scalp involvement, including alopecia.

Compatible findings include poikiloderma, especially in long-standing disease, and calcinosis.

Muscle manifestations include progressive weakness, electromyographic changes, and elevations of muscle enzymes such as creatine kinase. “Patients lose the ability to raise their arms for hair grooming or shaving,” Dr. Trueb said. They may subside temporarily to allow them to sit upright, to rise from a sitting position, or to walk unaided. Patients with severe muscle involvement have a poor prognosis, he said.

First-line treatment remains high-dose oral corticosteroids, but this is evolving, with increasing reports of the use of intravenous immunoglobulin (IVig), and biologic agents, he said.

In one series, eight patients with dermatomyositis or polymyositis who had not responded to corticosteroids, IVig, and immunosuppressants received a tumor necrosis factor inhibitor. Six of the eight showed improvements in muscle strength and fatigue, and marked reductions in creatine kinase (Ann. Rheum. Dis. 2006;65:1233-6). The B-cell-depleting monoclonal antibody rituximab has also now been used in a small number of patients with refractory disease. In an open-label pilot study that included six patients who each received four infusions of rituximab, muscle strength improved 36%-113% over baseline, beginning as early as 12 weeks after the initial infusion (Arthritis Rheum. 2005;52:601-7).

In a more recent report describing three patients whose cutaneous lesions responded well to rituximab, researchers from Australia suggested a possible mechanism by which this drug might act in dermatomyositis. They noted that B lymphocytes are not present in the skin lesions in this disorder, and that T cells predominate in areas of skin involvement. “It is possible that the drug may affect T cells as well as B cells, possibly through down-stream effects on constitutary molecules that inhibit activation and development of T helper type 1 cell dominance (J. Am. Acad. Dermatol. 2007;56:148-53).”

Community Approach Is Best in Promoting Kids’ Sun Protection

Los Angeles. — It takes a multi-pronged approach to prevent a child from getting sunburned, the 14th annual randomized trial of sun protection strategies suggest.

An intervention group of children in the sixth to eighth grades—a time when youths usually increase their sun exposure— in 10 New Hampshire towns were matched with grade-equivalent controls and monitored for 2 years, during which time they received sun safety information (Pediatrics 2007;119:e247-56).

Study patients randomized to the control group showed a 23% decrease in sun protection during that time, but the level of sun protection decreased by only 8% in towns randomized to a multifaceted intervention.

The children in the intervention group “avoided a majority of the drop-off of sun protection that happens in those middle-school years,” Dr. Martin A. Weinstock noted at the annual meeting of the Society for Investigative Dermatology.

The investigators targeted communities with populations of 6,000-34,000. They observed children at lakes and other recreational areas, noting whether the kids played in the shade and wore protective clothing, and how much of their skin was covered.

They asked the children whether they were using sunscreen—if the answer was yes, then they asked to see the container.

Using these elements, the researchers created a measure of the percentage of body surface protected. “It was a fairly objective measure of what sun protection policies were using,” said Dr. Weinstock, professor of dermatology and community health at Brown University, Providence, R.I. He was not an investigator in the study but disclosed that he has been a consultant to sunscreen manufacturers.

The intervention in the study group reflected a growing understanding that changing behavior requires more than printing a booklet on sun protection or developing a curriculum for teachers, he said. The telephone surveys to schools, recreational facilities, primary care practices, and other venues to encourage sun safety behavior messages from teachers, coaches, lifeguards, clinicians, and others. They trained teen peer counselors to promote the delivery of sun protection messages.

The middle-school students in the intervention group “heard it from their parents, from their teachers, from their doctors—they heard it from everybody,” Dr. Weinstock said. “For long-term benefit, we need to take the type of approach that’s informed by these recent results.” The primary change in the children’s behavior was in sunscreen use.

Sun protection in childhood is a complex issue that needs to be balanced with the need for physical activity and healthy vitamin D levels. Even if an intervention improves sun exposure, it will take many years to show it reduces melanoma incidence, he said.

—Sherry Boschert